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Retroperitoneal Cystic Lymphangioma

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Two cases of retroperitoneal cystic lymphangioma (CL) are presented; the current literature on this rare, benign neoplasm of the lymphatic system is reviewed. This tumor consists of various numbers of cyst-like cavities filled with a serous, serosanguineous or chylous fluid. The histogenesis of CL is still uncertain. Most commonly CL occurs in the neck and in the axillary region, whereas it is rare in the retroperitoneum. Although retroperitoneal CL is a benign lesion, it may cause significant morbidity due to its large size, and its often invasive character with a strong tendency to secondary infection. The treatment of choice is surgical excision.

KEY WORDS: **retroperitoneum, mesenchymal neoplasm, surgical excision**

INTRODUCTION

Retroperitoneal cystic lymphangiomas (CL) are rare tumors [1] usually found incidentally during diagnostic procedures performed for unrelated clinical reasons or at surgery [2]. When clinically significant, they generally present as a large, palpable mass causing displacement of abdominal organs; more rarely they can cause bowel or ureteral obstruction [3]. About 90% of the CL are diagnosed during the second year of life [4]; they rarely occur in adults. We report two cases of retroperitoneal CL, found in adult patients, successfully treated by surgical excision.

CASE REPORTS

Case 1

An intra-abdominal cystic mass was incidentally found in a 34-year-old man during an abdominal ultrasound performed for recurrent abdominal pain. A computed tomographic (CT) scan further defined its dimensions (25 cm in diameter) and retroperitoneal location (Fig. 1). The preoperative diagnosis was retroperitoneal tumor, possibly a cystic teratoma. At laparotomy a thin-walled large cystic mass was found involving the duodenum; a normal displaced pancreas was found. The mass appeared compatible with CL. Total excision of the mass required duodenal resection. The tumor contained chylous fluid. The pathologic diagnosis was cystic and cavernous lymphangioma. The postoperative course was uneventful. An 18-month follow-up disclosed no sign of recurrence.

Case 2

A 55-year-old man was operated on for a chronic and perforated duodenal ulcer. At laparotomy a cystic, soft, and whitish mass of 10 cm in diameter was incidentally found, displacing the hepatic pedicle and the inferior vena cava. The lesion was adherent to the pancreas, and its excision required distal pancreatic resection. The mass contained chylous fluid. The final diagnosis was CL. The postoperative course was uneventful. A 1-year follow-up disclosed no sign of recurrence.

PATHOLOGY

Grossly, CL are masses of thin-walled cyst-like cavities, often communicating each other. The cysts are filled with serous, serosanguineous or chylous fluid which contains varying amounts of protein and lipoid material [5]. The differences in the fluid depend on the degree of stasis and the number of channels that communicate with the lymphatic system [5].

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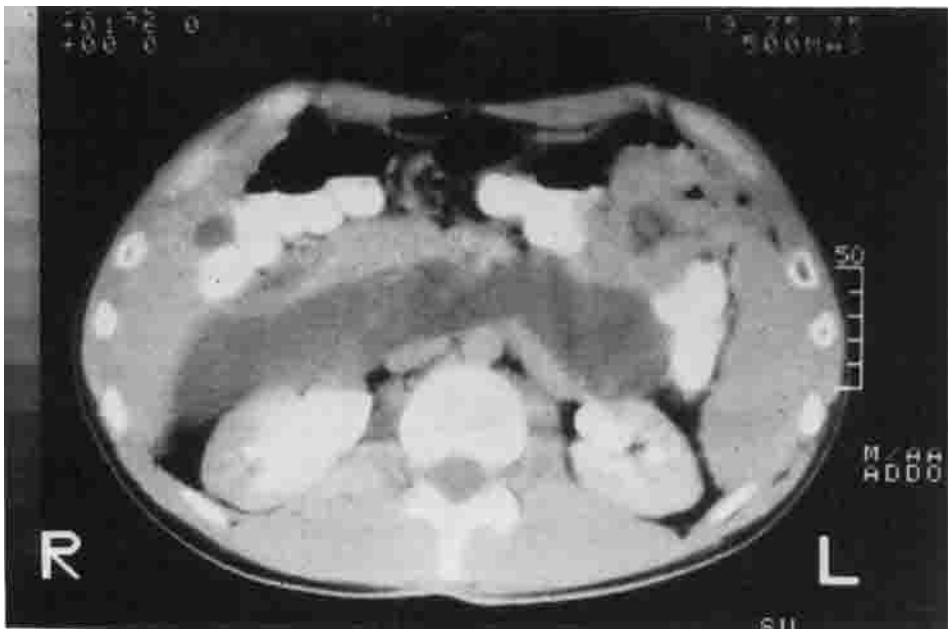

Fig. 1. The retroperitoneal position of a cystic and cavernous lymphangioma (Case 1) is shown. The CT scan disclosed pancreatic dislocation, inferior vena cava compression, and adhesion to the right vena cava. The left colon is anteriorly displaced.

Fig. 2. Cystic and cavernous lymphangioma: widely dilated, lymph-filled channels.

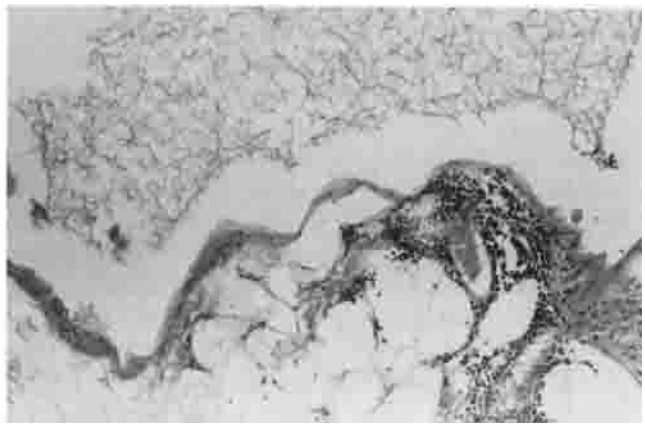

Fig. 3. Cystic and cavernous lymphangioma: proteinaceous material in lymphatic spaces and large collections of lymphocytes present in the stroma.

Microscopically, there are innumerable dilated lymphatic channels, often surrounded by lobules of adipose tissue [6] (Fig. 2). Cyst walls appear as a single layer of flattened endothelium [4] and contain a variable amount of fibrous and lymphoid tissue [6]. Endothelial proliferation is inconspicuous and the process is histologically benign [7].

Lymphangiomas have been classified as capillary (i.e., simple), cavernous, and cystic [8]. Only the cavernous and the cystic types have been observed in the retroperitoneum [8]. The capillary lymphangioma consists of an ill-defined mass composed of dilated lymph vessels with a richly cellular stroma. It is a compressible growth with a moderate number of channels in connection with the normal adjacent lymphatic system. Cavernous lymphangioma is a spongy, compressible tumor with dilated lymph vessels in an actively growing lymphoid stroma (Fig. 3). There is a moderate number of channels in connection with the normal adjacent lymphatic system.

It is noteworthy that only four cases of retroperitoneal cavernous lymphangioma had been reported in the literature [8], this case (Case 1) being the fifth as far as we know. CL is composed of one or multiple varying sized

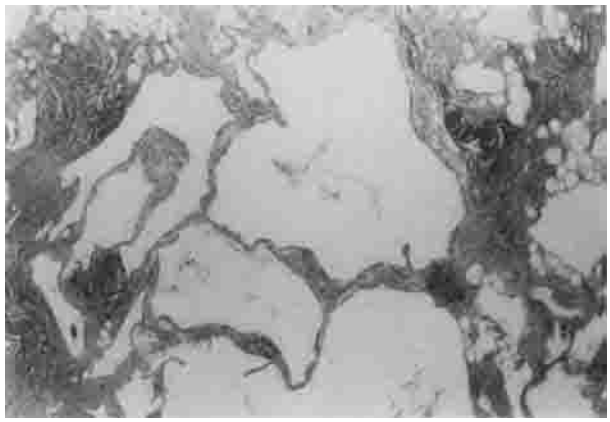

Fig. 4. CL: lymphatic spaces show gross cystic dilatation.

cysts with nearly no connections with normal adjacent lymphatics (Fig. 4). The cysts contain serous or chylous fluid.

The histogenesis of lymphangiomas is still uncertain. Various authors favor an acquired origin, due to obstruction of chylous vessels after inflammation, trauma, or degeneration. Others favor a congenital origin, due to proliferation of remnants of the embryonic lymph sac, for unknown reasons [8]. The most widely accepted theory is that of early developmental sequestration of lymphatic vessels that fail to establish connection with normal draining lymphatics and become markedly dilated under the pressure of accumulating lymph [4]. This hypothesis does not account, however, for the often invasive character of CL; in addition, an active growth of endothelium may exist, resulting in slow dissection through normal tissues and pressure atrophy of adjacent structures [9]: It is also possible that the nature of tissue surrounding a lymphangioma may determine its type. Muscle fibers may limit expansion of lymphatic cysts and the hamartoma remains cavernous, while a lesion located in fatty tissue or along fascial planes assumes a cystic form [10].

DISCUSSION

Many authors characterize CL as an abnormality of infancy and childhood [3,11], whereas the results of other studies [2,12–15] indicate that the CL, when located in the retroperitoneum, may occur in patients of all ages, and of both sexes [4].

Retroperitoneal CL can be found incidentally during an ultrasound or a CT scan performed for another clinical reason or at surgery. When clinically significant they generally present as a palpable mass that cannot be differentiated from other, more common retroperitoneal tumors. They can cause displacement or obstruction of the bowel and of the urinary tract [3]. Abdominal pain and fever, eventually with leukocytosis, may be present, probably related to hemorrhage or inflammation of the cystic wall [16–18].

The most characteristic radiologic finding of the retroperitoneal lymphangioma is a large tumor containing uncomplicated fluid with or without septa [12]. These features permit accurate discrimination from other retroperitoneal tumors [12]; cystic teratomas or abcesses, however, may produce similar findings and should be included in the differential diagnosis [7]. In the case of retroperitoneal CL reported here (Case 1), ultrasonography and CT led to the correct preoperative diagnosis.

Although retroperitoneal CL is a benign lesion, it may cause significant morbidity due to its large size, causing obstruction or dislocation of the bowel or urinary tract; it has a strong tendency to secondary infection [19] and invasion of surrounding structures. Malignant degeneration is rare [20].

As only rare cases showed spontaneous regression [19], the treatment of choice for retroperitoneal CL is surgical excision [21]. Prognosis is excellent [22]. The benign nature of the lesion warrants a conservative surgical approach. Sclerosing agents and radiotherapy have been employed in the past but should not be used as alternatives to surgery. In fact, it should be emphasized that there are cases of malignant transformation of previously irradiated lymphangiomas [19].

REFERENCES

1. Rauch RF: Retroperitoneal lymphangioma. Arch Surg 78:45–50, 1959.
2. Radin R, Weiner S, Koenigsberg M, et al.: Retroperitoneal cystic lymphangioma. AJR 140:733–734, 1983.
3. Singh S, Baboo ML, Pathak IC: Cystic lymphangioma in children: Report of 32 cases including lesions at rare sites. Surgery 69:947–951, 1971.
4. Rekhi BM, Esselstyn CB, Levy I, Mercer RD: Retroperitoneal cystic lymphangioma. Report of two cases and review of the literature. Cleve Clin Q 39:125–128, 1972.
5. Harrow BR: Retroperitoneal lymphatic cyst (cystic lymphangioma). J Urol 77:82–89, 1957.
6. Kissane JW: "Pathology of Infancy and Childhood, 2nd ed." St. Louis: Mosby, 1975, p 1161.
7. Leonidas JC, Brill PW, Bhan I, Smith TH: Cystic retroperitoneal lymphangioma in infants and children. Radiology 127:203–208, 1978.
8. Koshy A, Tandon RK, Kapur BML, et al.: Retroperitoneal lymphangioma. Am J Gastroenterol 69:485–490, 1978.
9. Gray SW, Skandalakis JE: "Embryology for Surgeons." Philadelphia: Saunders, 1972; pp 701–707.
10. Bill AH Jr, Sumner DS: A unified concept of lymphangioma and cystic hygroma. Surg Gynecol Obstet 120:79–86, 1965.
11. Harkins GA, Sabiston DC: Lymphangioma in infancy and childhood. Pediatr Surg 47:811–822, 1960.
12. Davidson AJ, Hartman DS: Lymphangioma of the retroperitoneum: CT and sonographic characteristics. Radiology 175:507–510, 1990.
13. Rossi L, Mandrioli R, Rossi A, Ugolotti U: Retroperitoneal cystic lymphangioma. Br J Radiol 55:676–678, 1982.
14. Castellino RA, Finkelstein S: Lymphographic demonstration of a retroperitoneal lymphangioma. Radiology 115:355–356, 1975.
15. Sarno RC, Carter BL, Bankoff MS: Cystic lymphangiomas: CT diagnosis and thin needle aspiration. Br J Radiol 57:424–426, 1984.

16. Barnett LA: Retroperitoneal cystic lymphangioma. JAMA 173:1111–1116, 1960.
17. Kalish M, Dorr R, Hoskins P: Retroperitoneal cystic lymphangioma. Urology 6:503–506, 1975.
18. Kelalis PP, King LR, Belman AB: "Clinical Pediatric Urology, Vol 2." Philadelphia: Saunders, 1976, p 991.
19. Enzinger FM, Weiss SW: Tumors of lymph vessels. In: Gay SM (ed): "Soft Tissue Tumors." St. Louis: Mosby, 1988, pp 614–637.
20. Walker AR, Putnam TC: Omental, mesenteric and retroperitoneal cysts - A clinical study of 33 new cases. Ann Surg 178:13–19, 1973.
21. Vanek VW, Philips AK: Retroperitoneal, mesenteric, and omental cysts. Arch Surg 119:838–842, 1984.
22. Hebra A, Brown MF, McGeehin KM, Ross AJ: Mesenteric, omental, and retroperitoneal cysts in children. South Med J 86(2):173–176, 1993.